Echinococcosis – a rare spinal disorder

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A 22-year-old woman presented with a 2-year history of lower back pain radiating to the right foot. On examination there were neurological deficits mainly of the right lower leg and foot. She had bladder and bowel involvement.

Blood tests showed a mild peripheral eosinophilia. Cysticercosis, hydatid and schistosomiasis complement fixation tests were all negative. The abdominal sonar was normal.

A magnetic resonance imaging (MRI) scan of the lumbar spine (Figs 1 and 2) demonstrated the presence of multiple cysts in the intraosseous, extradural and intrathecal compartments.

These cysts were removed using the laminectomy approach. More than 50 cysts of widely varying sizes were extracted (Fig. 3). Histological analysis showed these to be hydatid cysts.

Discussion

Hydatid disease caused by the parasite *Echinococcus granulosus* is a serious problem in much of Africa and is endemic in much of southern Africa. The Turkana people of northern Kenya have among the highest incidence rates of this disease in the world, owing to their nomadic, livestock-keeping lifestyle, reliance on hunting dogs and non-burial of their deceased.¹
The life cycle of *E. granulosus* involves mainly dogs as the definitive host, but also hyena and jackals. The intermediate hosts are mostly sheep, camel, cattle and goats, with humans becoming involved where there is contamination of the food and water supply with faecal matter from affected dogs.

Up to 70% of hydatid cysts occur in the liver and between 12% and 30% are found in the lungs. Bone involvement in hydatid disease has been reported to be only 0.5 - 4%, and spinal involvement is found in 50% of these cases with mortality of 18 - 50%.

Primary spinal involvement without evidence of disease elsewhere in the body, particularly the liver, has been considered atypical. This is because the portal venous system draining from the gut to the liver is the usual course of travel for these parasites. To explain this it is considered that the embryos may have penetrated the intestinal muscle and may have entered into the inferior vena cava system directly through small venous connections between this system and the portal circulation. Various conditions in daily life associated with the Valsava manoeuvre might have caused a movement of the embryos in the inferior vena cava system towards the retroperitoneum and spinal/paraspinal structures via the lumbar epidural venous plexus.

Clinically, patients may present in varied ways including with radiculopathy, myelopathy and local pain owing to bony destructive lesions and pathological fractures.

The treatment of hydatid disease of the spine is fraught with difficulties as there may be multiple cysts of all sizes in a single patient, with some deeply embedded in bone. It is important that all the cysts are removed, but unless this is painstakingly done there is frequent recurrence. A 100% recurrence rate has been reported in 11 patients followed up over an average of 20 years (range 1 - 34 years), each of whom had an average of 4.8 repeat surgical procedures to deal with these recurrences. Where the cysts are limited to the intradural compartment, this is less of a problem.

A second problem is dealing with intraoperative spillage of cyst contents, which contain protoscolices and can disseminate in tissue and grow new cysts. Although ideal, it is rarely possible to remove the cysts intact in every case. Scolicidal solutions have been suggested such as 5 - 10% formalin, 1% aqueous iodine, 0.5% silver nitrate or hypertonic saline to irrigate the area of spillage in order to kill off the scolices. None of these represent the results of class-one evidence trials, and are merely the recommendations of individual surgeons. Patients may suffer acute anaphylactic reactions to spilled cyst fluid and adrenaline, histamine and corticosteroids must be available to deal with these reactions.

Adjutant medical therapy presents another problem. Long-term treatment with albendazole has been the mainstay of treatment; however there is increasing evidence that it is less effective against bone cysts than cysts in the liver or lungs. The addition of praziquantel to albendazole therapy reduces the risk of postoperative recurrence but there is uncertainty about how long this therapy should be continued.

Extensive reconstruction and fusion procedures may be called for because of bone destruction. Poly-methylmethacrylate (PMMA) has been suggested as a way of restoring bone strength to obviate more costly procedures.

Strict follow-up is critical in the management of these patients. In order to ensure that any recurrences are detected early, ideally MRI scans should be done frequently during the postoperative period. This is expensive and out of the reach of many centres. Blood tests are also difficult to rely on in this regard.

Spinal hydatid disease should be included in the differential diagnosis when a patient presents with cysts in the spine or vertebral column.